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Technical Note

Pediatric peri-medullary arteriovenous fistula: Pearls for diagnosis and treatment

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ABSTRACT

Pediatric type IVc perimedullary arteriovenous fistulae (PAVF) comprise a rare subcategory of spinal vascular malformations in which multiple spinal arteries directly connect with draining veins resulting in high-flow arteriovenous shunting and large intradural venous varicosities. Complete disconnection of the fistula is necessary to prevent hemorrhage or spinal compression. A surgical, rather than endovascular, approach proves favorable under specific circumstances. Due to the rarity of these pediatric fistulae, no large studies exist to enumerate these circumstances. This case report fills this void by detailing several considerations which favored surgery for a type IVc PAVF in a 17-year-old female patient.

1. Introduction

First described by Djindjian et al. in 1977 and classified as a type IV spinal arteriovenous malformation (SAVM) by Heros et al. in 1986, perimedullary arteriovenous fistulae (PAVF) remain a rare entity in reported neurosurgical operative literature.[1] PAVFs are thoracolumbar vascular malformations arising commonly from the anterior spinal artery.[2] However, several case reports have recorded variant arterial origins including the intercostal and posterior spinal arteries. Type IV PAVFs are subcategorized according to the Anson-Spetzler convention based on angioarchitecture into 1) Type IVa which shows slow shunt flow without arterial or venous dilatation, 2) Type IVb which demonstrates slightly greater shunt flow with venous dilatation, and 3) Type IVc which arises from multiple arterial feeders and shows markedly increased shunt flow leading to tortuous intradural venous varices.[1]

While estimates suggest that type IV AVMs altogether comprise 17–39 % of all SAVMs, cases of type IVc PAVFs are largely limited to the pediatric population.[2] Type IVb and IVc PAVFs present with a range of acute neurological symptoms and subarachnoid hemorrhage (SAH) or hematomyelia. Concomitant vascular syndromes without a family history of PAVFs are also seen in up to 25 % pediatric patients with type IVb and IVc PAVFs.[1]

First-pass non-invasive imaging techniques such as 3D T2-weight magnetic resonance imaging (3D-T2W MRI) or computed tomographic angiography (CTA) are used to localize SAVMs.[3] Definitive diagnosis requires detailed imaging of angioarchitecture with digital subtraction angiography (DSA) to determine arterialization, venous draining, and shunt point(s).[1]

Current evidence regarding pediatric type IVc PAVFs is limited to case reports due to their rarity. Published reports recommend endovascular embolization as the first option of treatment.[3,4] In instances of anticipated difficulties in catheterization due to hazardous location or tortuosity of feeder vessels, surgery should be considered; nevertheless, very few surgical cases have been reported. Herein, we report a case of an adolescent female presenting with SAH requiring surgical ligation of a type IVc PAVF.

2. Patient information and clinical findings

A 17-year-old female patient with no significant past medical history presented to our institution with severe refractory headache, vomiting without nausea, and left-sided weakness. The patient was grossly neurologically intact except for a mild left pronator drift and headaches.

Abbreviations: Indocyanine green, ICG; perimedullary arteriovenous fistula, PAVF; spinal arteriovenous malformation, SAVM.

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3. Diagnostic assessment

Non-contrast head CT demonstrated SAH with intraventricular hemorrhage (IVH) without hydrocephalus. Diagnostic cerebral angiography revealed no intracranial vascular pathology. In reviewing the initial CTA Head/Neck, suspicious enlarged vasculature noted in the posterior cervical spinal canal, raised the concern for enlarged venous structures [Fig. 1A]. MRI of the spine confirmed high suspicion for spinal vascular malformation [Fig. 1B]. Diagnostic spinal angiography elucidated the underlying pathology as a Type IV PAVF with arterial supply from the right supreme intercostal and T5 radicular arteries [Fig. 2A].

4. Therapeutic intervention

This case was discussed in our multidisciplinary conference with NeuroInterventional Radiology and Vascular Neurosurgery. Given the patient's young age, the acute history of rupture, the suboptimal visualization of the fistulous point due to large venous varices, and the tortuosity of primary feeding intercostal branch from the thyrocervical trunk, it was agreed to proceed with primary surgical ligation of the fistula. The patient was prepped and taken to the operating room the following day for a T2-4 laminoplasty and surgical ligation of the PAVF (Supplemental video). During surgical consent, we obtained consent from the patient for use of case details, images, and videos for educational purposes.

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On the day of the procedure, a standard midline T2-T4 laminotomy was performed, with intraoperative neurophysiologic monitoring and C-arm fluoroscopy. A midline durotomy was performed and blunt dissection of the arachnoid layer revealed the underlying serpiginous arteriovenous complex. The descending perimedullary posterior feeding artery seen on angiography was identified and followed distally toward the fistulous point proximal to the venous varices of the AVF. A loop of the vessel was noted proximal to the fistulous point; this loop contained an additional branch concerning a secondary feeding vessel. Arterialized draining of the AVF was visualized with indocyanine green (ICG) angiography to confirm early venous filling and reveal the fistulous point. A temporary mini aneurysm clip was placed at this suspected point, after which motor and sensory monitoring revealed no changes from baseline. The temporary clip was removed, and two, 2 mm AVM clips were placed across the fistulous point. The communicating vessel was coagulated using bipolar electrocautery and ligated. Repeat ICG showed persistent filling of the venous varices which seemed to arise from the additional branch noted at the distal loop of the vessel.

Needle Doppler was performed on this secondary vessel and identified arterial flow. Another temporary clip was placed, and neuromonitoring reported a decrease in motor-evoked potentials along the right lower extremity. Further dissection of the secondary vessel was performed, and a more distal location was temporarily clipped. Neuromonitoring reported no changes from baseline. Another ICG showed persistent filling of the venous varices, so another temporary clip was placed further distally. Repeat ICG revealed delayed filling of the varices with decreased signal consistent with stasis. Neuromonitoring also reported no changes from baseline at this more distal location. The secondary vessel was cauterized at both temporary clip locations using bipolar electrocautery and ligated. Following this ligation, a color change was noted in the venous varices, suggesting reduced arterial flow. Final neuromonitoring confirmed no changes relative to baseline.

5. Follow-up and outcomes

On hospital day 17, the patient was discharged without any neurologic deficits. Postsurgical diagnostic spinal angiography [Fig. 2B] at this time confirmed exclusion of the spinal dural AVF. At a six-month follow-up visit, the patient remained neurologically intact without any new symptoms or concerns. MRI at this six-month visit [Fig. 1C] confirmed sustained exclusion of the previously observed large vascular malformation.

6. Discussion

The patient in this case report presented with symptoms and imaging findings consistent with SAH related to a Type IVc PAVF. Acute bleeding, which carries a 20 % mortality rate if untreated,[5] is the most common complication of these rare spinal fistulas. They exhibit a 2.55 % annual rupture risk rate, and the lifetime hemorrhage likelihood ranges from 50 % to 70 % without intervention.[3] Chronic symptoms, while less common, can carry incredible morbidity including bowel or bladder incontinence, radiculopathy, or progressive paraplegia.[6] Prompt intervention, however, both protects against future deficits and helps to improve existing symptoms.[1,3,6]

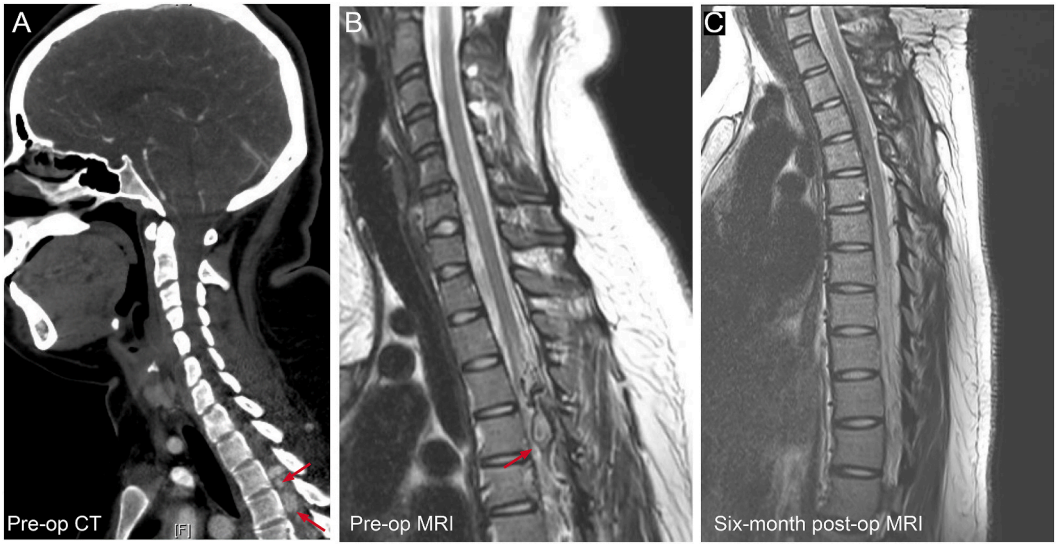

Fig. 1. Perioperative diagnostic imaging (A) Pre-operative sagittal CTA head and neck showing enlarged vessels in the upper thoracic spinal canal (red arrows) (B) Pre-operative sagittal MRI showing suspected vascular malformation at the level of T2-T4 (red arrow) (C) Six-month post-operative Sagittal MRI showing sustained exclusion of the vascular malformations.

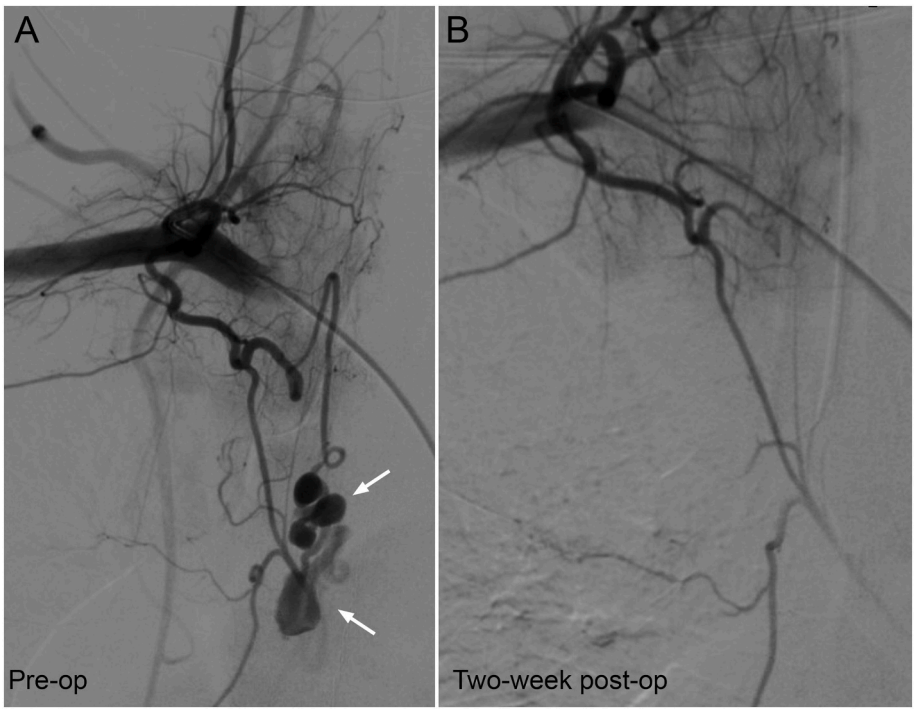

Fig. 2. Perioperative diagnostic spinal angiograms. Right costocervical trunk injection, AP view (A) Pre-operative run demonstrates Type 4c spinal AV fistula supplied by right supreme intercostal artery arising from costocervical trunk. Noted is collateral supply from the right T5 intercostal artery. Perimedullary artery noted to make a hairpin turn at superior margin of abnormality before reaching fistulous point obscured on image by multiple venous varices (arrows) (B) Two-week post-operative run depicting the supreme intercostal artery with no intradural continuation and no residual filling of previously seen perimedullary AV fistula. Still seen is right T5 intercostal collateralization.

By reviewing several case reports of these rare pediatric spinal AVFs,[4,7–10] it became apparent that these Type III/IV AVMs most often present with sensory and motor disturbances in the lower limbs with more variable systemic and urinary symptoms [Table 1]. In all but one case, lasting symptom remission was achieved with either endovascular or surgical intervention. Though an endovascular approach may be favorable when treating many spinal cord AVFs,[3,4] surgical ligation is preferred under certain circumstances.[5,6] Due to the rarity of PAVFs, especially pediatric fistulas, no large-scale, multi-institution studies exist to guide treatment. For this reason, case reports like ours are vital to informing clinical decision-making.

Our patient's case reveals several anatomical complexities which favor surgical intervention. First, the dorsal rather than the ventral location of the fistula meant an open approach was more feasible. Second, imaging evidence pointed to multiple feeders making disconnection at the fistulous point the key to successful treatment. Third, large venous varices obscured the fistulous point from visualization on angiogram and therefore hampered its accessibility. Finally, marked tortuosity of the feeding artery meant advancing an endovascular catheter could prove difficult.

In addition to lessons in clinical decision-making, this case also demonstrated the paramount importance of intraoperative indocyanine green (ICG) dye. Our pre-operative suspicion of multiple arterial feeders was validated by the unsuccessful disconnection of the fistula from arterial supply following the ligation of the first vessel. The subsequent use of ICG enabled identification of the second feeding artery and better visualization of the fistulous point. After ligating the second vessel, a final ICG application confirmed the elimination of the arterio-venous shunting. This case demonstrated that aggressive use of ICG can make successful surgical ligation of these fistulas more likely.

Table 1
Literature review of case reports describing the management of pediatric spinal AVMs.

Authors	Management	Patient Information	Clinical Presentation	Spinal AVM type and location	Follow up outcomes
Current case	Surgical	17 year-old F; no significant PMH	Sudden onset headache, vomiting, and left-sided weakness	Type IVc AVM; perimedullary AVF; T2/4	Neurologically intact and symptom free after 6 months
Bertoli et al., 2021[7]	Surgical	15 month-old F; PMH congenital syphilis	1 week fever, urinary retention, and 1 day of paraplegia	Type IVc AVM; perimedullary AVF; L1/L2	Refractory paraplegia and neurogenic bladder despite sustained AVF exclusion after 1 year
Chen et al., 2008[8]	Surgical	8 year-old M; no significant PMH	Sudden upper back and right chest wall pain, lower limb weakness, urinary incontinence	Type III AVM; intramedullary AVF; T1/2	Neurologically intact and symptom free after 6 months
Chu et al., 2022[9]	Endovascular	16 year-old M; no significant PMH	3 months progressive lower limb numbness and weakness	Type IV AVM; perimedullary AVF; L2	Resolution of symptoms at post-operative discharge
Bankole et al., 2021[4]	Endovascular	12 year-old F; no significant PMH	1 year back and leg pain with sudden onset right leg paresis and gait disorder	Type IV AVM; perimedullary AVF; T10	Neurologically intact and symptom free after 9 months
Rajadurai et al., 2020[10]	Endovascular	2 year-old F; no significant PMH	Sudden onset lethargy, gait ataxia, and left lower limb paresis	Type IV AVM; perimedullary AVF; T3/4	Neurologically intact and symptom free after 2 years

Female (F); Male (M); Past medical history (PMH); Arteriovenous malformation (AVM); Arteriovenous fistula (AVF).

7. Conclusion

The clinical course of the presented patient demonstrates careful evaluation and microsurgical technique to achieve complete ligation of an anatomically complex type IVc PAVF.

CRediT authorship contribution statement

Joshua A. Reynolds: Writing – review & editing, Writing – original draft, Formal analysis, Conceptualization. **Yashraj Srivastava:** Writing – review & editing, Writing – original draft, Conceptualization. **Muhammed Amir Essibayi:** Writing – review & editing, Writing – original draft, Conceptualization. **Anna Nia:** Writing – review & editing, Writing – original draft, Data curation, Conceptualization. **Adisson Fortunel:** Writing – review & editing, Writing – original draft, Methodology, Data curation, Conceptualization. **Neil Haranhalli:** Writing – review & editing, Writing – original draft, Supervision, Methodology, Data curation, Conceptualization.

Declaration of competing interest

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.wnsx.2024.100401.

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Abbreviations

3D-T2W MRI: 3D T2-weight magnetic resonance imaging
CTA: computed tomographic angiography
DSA: digital subtraction angiography
ICG: Indocyanine green
IVH: intraventricular hemorrhage
PAVF: perimedullary arteriovenous fistula
SAH: subarachnoid hemorrhage
SAVM: spinal arteriovenous malformation